Complete Summary

GUIDELINE TITLE

Pulmonary hypertension/Eisenmenger physiology. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease).

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Pulmonary hypertension/Eisenmenger physiology. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 2008;52(23):e210-15.

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Adult congenital heart disease
- Pulmonary hypertension
- Eisenmenger syndrome

GUIDELINE CATEGORY

Counseling
Diagnosis
Evaluation
Management
Risk Assessment
Treatment

CLINICAL SPECIALTY

Cardiology
Family Practice
Internal Medicine
Obstetrics and Gynecology
Radiology
Thoracic Surgery

INTENDED USERS

Health Care Providers
Physicians

GUIDELINE OBJECTIVE(S)

- To assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions associated with adult congenital heart disease (ACHD)
- To define practices that meet the needs of most patients in most circumstances
- To support the practicing cardiologist in the care of ACHD patients by providing a consensus document that outlines the most important diagnostic and management strategies and indicates when referral to a highly specialized center is appropriate

TARGET POPULATION

Adults with congenital heart disease, pulmonary hypertension and/or Eisenmenger syndrome

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

Noninvasive assessment of cardiovascular anatomy and potential shunting

- Pulse oximetry
- Chest x-ray
- Electrocardiogram
- Transesophageal echocardiography
- Transthoracic echocardiography
- Magnetic resonance imaging (MRI)
- Computed tomography (CT)
- Complete blood count
- Nuclear lung scintigraphy
- Cardiac catheterization
- Pulmonary function tests
- Pulmonary embolism-protocol CT with parenchymal lung windows
- Six minute walk test

Management/Treatment

1. Prompt therapy for arrhythmias and infections
2. Yearly testing
    - Hemoglobin
    - Platelet count
    - Iron stores
    - Creatinine
    - Uric acid
3. Pulmonary vasodilator therapy
4. Rigorous medication review
5. Counseling of patients to avoid high-risk activities and exposures
6. Reproductive counseling, including counseling to avoid pregnancy and estrogen-containing contraceptives
7. Follow-up
    - Coordinated care under the supervision of a trained congenital heart disease and pulmonary arterial hypertension provider
    - Annual comprehensive evaluation

MAJOR OUTCOMES CONSIDERED

- Sudden Cardiac Death
- Cardiac Arrest
- Mortality
- Heart and Heart/Lung Transplantation

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Unlike other American College of Cardiology/American Heart Association (ACC/AHA) practice guidelines; there is not a large body of peer-reviewed published evidence to support most recommendations, which will be clearly indicated in the text. An extensive literature survey was conducted that led to the incorporation of 647 references. Searches were limited to studies, reviews, and other evidence conducted in human subjects and published in English. Key search words included but were not limited to adult congenital heart disease (ACHD), atrial septal defect, arterial switch operation, bradycardia, cardiac catheterization, cardiac reoperation, coarctation, coronary artery abnormalities, cyanotic congenital heart disease, Doppler-echocardiography, d-transposition of the great arteries, Ebstein's anomaly, Eisenmenger physiology, familial, heart defect, medical therapy, patent ductus arteriosus, physical activity, pregnancy, psychosocial, pulmonary arterial hypertension, right heart obstruction, supravalvular pulmonary stenosis, surgical therapy, tachyarrhythmia, tachycardia, tetralogy of Fallot, transplantation, tricuspid atresia, and Wolff-Parkinson-White. Additionally, the writing committee reviewed documents related to the subject matter previously published by the ACC and AHA. References selected and published in this document are representative and not all-inclusive.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFEC		
		CLASS I *Benefit >>> Risk* Procedure/Treatment **SHOULD** be performed/administered	**CLASS IIa** *Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	**CLASS IIb** *Benefit ≥ Risk* *Additional stud* *objectives nee* *registry data w* *helpful* Procedure/Trea **MAY BE CONS**
Estimate of Certainty (Precision) of Treatment	**LEVEL A** Multiple population evaluated*	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from	• Recomm usefuln less we • Greater evidenc multiple

		SIZE OF TREATMENT EFFEC		
Effect	Data derived from multiple randomized clinical trials or meta-analyses	randomized trials or meta-analyses	multiple randomized trials or meta-analyses	trials or analyse
	LEVEL B Limited population evaluated* Data derived from a single randomized clinical trial or nonrandomized studies	• Recommendation that procedure or treatment is useful/effective • Evidence from single randomized trial or nonrandomized studies	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from single randomized trial or nonrandomized studies	• Recomm usefuln less we • Greater evidenc random nonran studies
	LEVEL C Very limited population evaluated* Only consensus opinion of experts, case studies or standard of care.	• Recommendation that procedure or treatment is useful/effective • Only expert opinion, case studies, or standard-of-care	• Recommendation in favor of treatment of procedure being useful/effective • Only diverging expert opinion, case studies, or standard-of-care	• Recomm usefuln less we • Only di opinion or stan

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The committee reviewed and ranked evidence supporting current recommendations with the weight of evidence ranked as Level A if the data were derived from multiple randomized clinical trials involving a large number of individuals. The committee ranked available evidence as Level B when data were derived from a limited number of trials involving a comparatively small number of patients or from well-designed data analyses of nonrandomized studies or observational data registries. Evidence was ranked as Level C when the consensus of experts was the primary source of the recommendation. In the narrative portions of these guidelines, evidence is generally presented in chronological order of development. Studies are identified as observational, randomized, prospective, or retrospective. The committee emphasizes that for certain conditions for which no other therapy is available, the indications are based on expert consensus and years of clinical experience and are thus well supported, even though the evidence was ranked as Level C. An analogous example is the use of penicillin in pneumococcal pneumonia where there are no randomized trials and only clinical experience. When indications at Level C are supported by historical clinical data, appropriate references (e.g., case reports and clinical reviews) are cited if available. When Level C indications are based strictly on committee consensus, no references are cited. The final recommendations for indications for a diagnostic procedure, a particular therapy, or an intervention in adult congenital heart disease (ACHD) patients summarize both clinical evidence and expert opinion. The schema for classification of recommendations and level of evidence illustrates how the grading system provides an estimate of the size of treatment effect and an estimate of the certainty of the treatment effect (see "Rating Scheme for the Strength of the Evidence" above).

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines was formed to create clinical practice guidelines for select cardiovascular conditions with important implications for public health. This guideline writing committee was assembled to adjudicate the evidence and construct recommendations regarding the diagnosis and treatment of adult congenital heart disease (ACHD). Writing committee members were selected with attention to ACHD subspecialties, broad geographic representation, and involvement in academic medicine and clinical practice. The writing committee included representatives of the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

Writing committees are specifically charged to perform a formal literature review, weigh the strength of evidence for or against particular treatments or procedures, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that might

influence the choice of tests or therapies are considered, as well as the frequency of follow-up and cost-effectiveness. When available, information from studies on cost is considered, but data on efficacy and clinical outcomes constitute the primary basis for recommendations in these guidelines.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

See "Rating Scheme for the Strength of the Evidence" field, above.

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

This document was reviewed by 3 external reviewers nominated from both the American College of Cardiology (ACC) and the American Heart Association (AHA), as well as reviewers from the American Society of Echocardiography, Canadian Cardiovascular Society, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, and Society of Thoracic Surgeons, and 20 individual content reviewers which included reviewers from the ACC Congenital Heart Disease and Pediatric Cardiology Committee and the AHA Congenital Cardiac Defects Committee. All reviewer relationships with industry information were collected and distributed to the writing committee and are published in the original guideline document (see the "Conflicts of Interest/Financial Disclosures" field in this document).

This document was approved for publication by the governing bodies of the American College of Cardiology Foundation (ACCF) and the AHA and endorsed by the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) classification of the recommendations for patient evaluation and treatment (classes I-III) and the levels of evidence (A-C) are defined at the end of the "Major Recommendations" field.

Recommendations for Evaluation of the Patient With Congenital Heart Disease–Pulmonary Arterial Hypertension

Class I

1. Care of adult patients with congenital heart disease (CHD)-related pulmonary arterial hypertension (PAH) should be performed in centers that have shared expertise and training in both adult congenital heart disease (ACHD) and PAH. (***Level of Evidence: C***)
2. The evaluation of all ACHD patients with suspected PAH should include noninvasive assessment of cardiovascular anatomy and potential shunting, as detailed below:
    a. Pulse oximetry, with and without administration of supplemental oxygen, as appropriate. (***Level of Evidence: C***)
    b. Chest x-ray. (***Level of Evidence: C***)
    c. Electrocardiogram (ECG). (***Level of Evidence: C***)
    d. Diagnostic cardiovascular imaging via transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), magnetic resonance imaging (MRI), or computed tomography (CT) as appropriate. (***Level of Evidence: C***)
    e. Complete blood count and nuclear lung scintigraphy. (***Level of Evidence: C***)
3. If PAH is identified but its causes are not fully recognized, additional testing should include the following:
    a. Pulmonary function tests with volumes and diffusion capacity (diffusing capacity of the lung for carbon monoxide). (***Level of Evidence: C***)
    b. Pulmonary embolism–protocol CT with parenchymal lung windows. (***Level of Evidence: C***)
    c. Additional testing as appropriate to rule out contributing causes of PAH. (***Level of Evidence: C***)
    d. Cardiac catheterization at least once, with potential for vasodilator testing or anatomic intervention, at a center with expertise in catheterization, PAH, and management of CHD-PAH. (***Level of Evidence: C***)

Class IIa

1. It is reasonable to include a 6-minute walk test or similar nonmaximal cardiopulmonary exercise test as part of the functional assessment of patients with CHD-PAH. (***Level of Evidence: C***)

Management Strategies

Recommendations for Medical Therapy of Eisenmenger Physiology

Class I

1. It is recommended that patients with Eisenmenger syndrome avoid the following activities or exposures, which carry increased risks:
    a. Pregnancy. (***Level of Evidence: B***)
    b. Dehydration. (***Level of Evidence: C***)
    c. Moderate and severe strenuous exercise, particularly isometric exercise. (***Level of Evidence: C***)
    d. Acute exposure to excessive heat (e.g., hot tub or sauna). (***Level of Evidence: C***)

    e. Chronic high-altitude exposure, because this causes further reduction in oxygen saturation and increased risk of altitude-related cardiopulmonary complications (particularly at an elevation greater than 5000 feet above sea level). (***Level of Evidence: C***)
    f. Iron deficiency. (***Level of Evidence: B***)
2. Patients with Eisenmenger syndrome should seek prompt therapy for arrhythmias and infections. (***Level of Evidence: C***)
3. Patients with Eisenmenger syndrome should have hemoglobin, platelet count, iron stores, creatinine, and uric acid assessed at least yearly. (***Level of Evidence: C***)
4. Patients with Eisenmenger syndrome should have assessment of digital oximetry, both with and without supplemental oxygen therapy, at least yearly. The presence of oxygen-responsive hypoxemia should be investigated further. (***Level of Evidence: C***)
5. Exclusion of air bubbles in intravenous tubing is recommended as essential during treatment of adults with Eisenmenger syndrome. (***Level of Evidence: C***)
6. Patients with Eisenmenger syndrome should undergo noncardiac surgery and cardiac catheterization only in centers with expertise in the care of such patients. In emergent or urgent situations in which transportation is not feasible, consultation with designated caregivers in centers with expertise in the care of patients with Eisenmenger syndrome should be performed and sustained throughout care. (***Level of Evidence: C***)

Class IIa

1. All medications given to patients with Eisenmenger physiology should undergo rigorous review for the potential to change systemic blood pressure, loading conditions, intravascular shunting, and renal or hepatic flow or function. (***Level of Evidence: C***)
2. Pulmonary vasodilator therapy can be beneficial for patients with Eisenmenger physiology because of the potential for improved quality of life. (***Level of Evidence: C***)

Key Issues to Evaluate and Follow-Up

Recommendations for Reproduction

Class I

1. Women with severe CHD-PAH, especially those with Eisenmenger physiology, and their partners should be counseled about the absolute avoidance of pregnancy in view of the high risk of maternal death, and they should be educated regarding safe and appropriate methods of contraception. (***Level of Evidence: B***)
2. Women with CHD-PAH who become pregnant should:
    a. Receive individualized counseling from cardiovascular and obstetric caregivers collaborating in care and with expertise in management of CHD-PAH. (***Level of Evidence: C***)
    b. Undergo the earliest possible pregnancy termination after such counseling. (***Level of Evidence: C***)

3. Surgical sterilization carries some operative risk for women with CHD-PAH but is a safer option than pregnancy. In view of advances in minimally invasive techniques, the risks and benefits of sterilization modalities should be discussed with an obstetrician experienced in management of high-risk patients, as well as with a cardiac anesthesiologist. (***Level of Evidence: C***)

Class IIb

1. Pregnancy termination in the last 2 trimesters of pregnancy poses a high risk to the mother. It may be reasonable, however, after the risks of termination are balanced against the risks of continuation of the pregnancy. (***Level of Evidence: C***)

Class III

1. Pregnancy in women with CHD-PAH, especially those with Eisenmenger physiology, is not recommended and should be absolutely avoided in view of the high risk of maternal mortality. (***Level of Evidence: B***)
2. The use of single-barrier contraception alone in women with CHD-PAH is not recommended owing to the frequency of failure. (***Level of Evidence: C***)
3. Estrogen-containing contraceptives should be avoided. (***Level of Evidence: C***)

Recommendations for Follow-Up

Class I

1. Patients with CHD-related PAH should:
    a. Have coordinated care under the supervision of a trained CHD and PAH provider and be seen by such individuals at least yearly. (***Level of Evidence: C***)
    b. Have yearly comprehensive evaluation of functional capacity and assessment of secondary complications. (***Level of Evidence: C***)
    c. Discuss all medication changes or planned interventions with their CHD-related PAH caregiver. (***Level of Evidence: C***)

Class III

1. Endocardial pacing is not recommended in patients with CHD-PAH with persistent intravascular shunting, and alternative access for pacing leads should be sought (the risks should be individualized). (Khairy et al., 2006) (***Level of Evidence: B***)

Definitions:

Applying Classification of Recommendations and Level of Evidence

Â	**SIZE OF TREATMENT EFFEC**		
Â	**CLASS I**	**CLASS IIa**	**CLASS IIb**

Â		SIZE OF TREATMENT EFFEC		
		Benefit >>> Risk Procedure/Treatment **SHOULD** be performed/administered	*Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	*Benefit ≥ Risk* *Additional stud objectives nee registry data w helpful* Procedure/Trea **MAY BE CONS**
Estimate of Certainty (Precision) of Treatment Effect	**LEVEL A** Multiple population evaluated* Data derived from multiple randomized clinical trials or meta-analyses	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses	• Recomm usefuln less we • Greater evidenc multiple trials or analyse
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*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply

that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

REFERENCES SUPPORTING THE RECOMMENDATIONS

References open in a new window

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate management of patients with congenital heart disease-related pulmonary hypertension and Eisenmenger physiology

POTENTIAL HARMS

- The theoretical possibility of worsening of right-to-left shunting raises questions about the safety of using pulmonary artery modulating therapies that also have systemic vasodilator potential. Nevertheless, some of these agents (intravenous prostacyclin and oral sildenafil) have yielded improvements in hemodynamics, exercise tolerance, and/or systemic arterial oxygen saturation in limited case studies. The potential for significant adverse reaction due to these agents has been recognized.
- Maternal sterilization carries a defined operative risk of mortality, and endoscopic sterilization may be the safer option. Hormonal therapies increase the preexisting potential for thrombosis, although progesterone-only preparations may be considered. Barrier methods have an increased rate of failure, and intrauterine device implantation carries anecdotally increased infection risk, although the highest risk is for local infection in multipartner couples. There is no consensus on comparative contraceptive risks; therefore,

the patient should discuss options with a high-risk obstetrician (maternal fetal medicine specialist).

CONTRAINDICATIONS

CONTRAINDICATIONS

- In adults with Eisenmenger physiology, recognition of in vivo pulmonary thrombus, contrasted with reports of in vitro abnormalities of coagulation in persons with cyanosis, has led to debate over the potential benefit of oral anticoagulant therapy, particularly with the concomitant bleeding diathesis inherent in the condition. In patients with active or chronic hemoptysis, anticoagulation is contraindicated.
- Pregnancy is contraindicated in women with congenital heart disease and pulmonary arterial hypertension (CHD-PAH).
- Estrogen-containing contraceptives should be avoided in women with CHD-PAH.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- These practice guidelines are intended to assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions. Clinicians should consider the quality and availability of expertise in the area where care is provided. These guidelines attempt to define practices that meet the needs of most patients in most circumstances. The recommendations reflect a consensus of expert opinion after a thorough review of the available current scientific evidence and are intended to improve patient care.
- Patient adherence to prescribed and agreed upon medical regimens and lifestyles is an important aspect of treatment. Prescribed courses of treatment in accordance with these recommendations are only effective if they are followed. Because lack of patient understanding and adherence may adversely affect outcomes, physicians and other healthcare providers should make every effort to engage the patient's active participation in prescribed medical regimens and lifestyles.
- If these guidelines are used as the basis for regulatory or payer decisions, the goal is quality of care and serving the patient's best interest. The ultimate judgment regarding care of a particular patient must be made by the healthcare provider and the patient in light of all of the circumstances presented by that patient. There are circumstances in which deviations from these guidelines are appropriate.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Slide Presentation

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Pulmonary hypertension/Eisenmenger physiology. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 2008;52(23):e210-15.

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008

GUIDELINE DEVELOPER(S)

American College of Cardiology Foundation - Medical Specialty Society
American Heart Association - Professional Association

SOURCE(S) OF FUNDING

The American College of Cardiology Foundation and the American Heart Association. No outside funding accepted.

GUIDELINE COMMITTEE

American College of Cardiology/American Heart Association Task Force on Practice Guidelines

Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Task Force Members: Sidney C. Smith, Jr, MD, FACC, FAHA, *Chair*; Alice K. Jacobs, MD, FACC, FAHA, *Vice-Chair*; Cynthia D. Adams, RSN, PhD, FAHA#; Jeffrey L. Anderson, MD, FACC, FAHA#; Elliott M. Antman, MD, FACC, FAHA**; Christopher E. Buller, MD, FACC; Mark A. Creager, MD, FACC, FAHA; Steven M. Ettinger, MD, FACC; Jonathan L. Halperin, MD, FACC, FAHA#; Sharon A. Hunt, MD, FACC, FAHA#; Harlan M. Krumholz, MD, FACC, FAHA; Frederick G. Kushner, MD, FACC, FAHA; Bruce W. Lytle, MD, FACC, FAHA#; Rick A. Nishimura, MD, FACC, FAHA; Richard L. Page, MD, FACC, FAHA; Barbara Riegel, DNSc, RN, FAHA#; Lynn G. Tarkington, RN; Clyde W. Yancy, MD, FACC, FAHA

Committee Members: Carole A. Warnes, MD, FRCP, FACC, FAHA, *Co-Chair*; Roberta G. Williams, MD, MACC, FAHA, *Co-Chair*; Thomas M. Bashore, MD, FACC; John S. Child, MD, FACC, FAHA; Heidi M. Connolly, MD, FACC; Joseph A. Dearani, MD, FACC*; Pedro del Nido, MD; James W. Fasules, MD, FACC; Thomas P. Graham, Jr, MD, FACC†; Ziyad M. Hijazi, MBBS, MPH, FACC, FSCAI‡; Sharon A. Hunt, MD, FACC, FAHA; Mary Etta King, MD, FACC, FASE§; Michael J. Landzberg, MD, FACC; Pamela D. Miner, RN, MN, NP; Martha J. Radford, MD, FACC; Edward P. Walsh, MD, FACC||; Gary D. Webb, MD, FACC¶

*Society of Thoracic Surgeons representative.

†International Society for Adult Congenital Heart Disease representative.

‡Society for Cardiovascular Angiography and Interventions representative.

§American Society of Echocardiography representative.

||Heart Rhythm Society representative.

¶Canadian Cardiovascular Society representative.

#Former Task Force member during this writing effort.

**Immediate past chair.

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines makes every effort to avoid actual, potential, or perceived conflicts of interest that might arise as a result of industry relationships or personal interests among the writing committee. Specifically, all members of

the writing committee, as well as peer reviewers of the document, are asked to disclose all such relationships that might be perceived as real or potential conflicts of interest. Writing committee members are also strongly encouraged to declare previous relationships with industry that might be perceived as relevant to guideline development. If a writing committee member develops a new relationship with industry during their tenure, they are required to notify guideline staff in writing. These statements are reviewed by the parent task force, reported orally to all members at each meeting of the writing committee, and updated and reviewed by the writing committee as changes occur.

Author Relationships With Industry and Other Entities–ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

Committee Member	Research Grant	Speakers' Bureau	Stock Ownership	Board of Directors	Consultant/Advisory Member
Dr. Carole A. Warnes (*Co-Chair*)	None	None	None	None	None
Dr. Roberta G. Williams (*Co-Chair*)	None	None	None	None	None
Dr. Thomas M. Bashore	None	None	None	None	None
Dr. John S. Child	None	None	None	None	None
Dr. Heidi M. Connolly	None	None	None	None	None
Dr. Joseph A. Dearani	None	None	None	None	None
Dr. Pedro del Nido	None	None	None	None	None
Dr. James W. Fasules	None	None	None	None	None
Dr. Thomas P. Graham, Jr	None	None	None	None	None
Dr. Ziyad M. Hijazi	None	None	None	None	• AGA Medical
Dr. Sharon A. Hunt	None	None	None	None	None

Committee Member	Research Grant	Speakers' Bureau	Stock Ownership	Board of Directors	Consultant/Advisory Member
Dr. Mary Etta King	None	None	None	None	None
Dr. Michael J. Landzberg	• Actelion • AGA • Medical • Myogen • NMT Medical • Pfizer	None	None	None	None
Dr. Pamela D. Miner	None	None	None	None	None
Dr. Martha J. Radford	None	None	None	None	None
Dr. Edward P. Walsh	None	None	None	None	None
Dr. Gary D. Webb	None	None	None	None	None

This table represents the relevant relationships of committee members with industry and other entities that were reported orally at the initial writing committee meeting and updated in conjunction with all meetings and conference calls of the writing committee during the document development process. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of $10,000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted.

See Appendix 2 in the original guideline document for peer reviewer relationships with industry.

ENDORSER(S)

American Society of Echocardiography - Professional Association
Heart Rhythm Society - Professional Association
International Society for Adult Congenital Heart Disease - Disease Specific Society
Society for Cardiovascular Angiography and Interventions - Medical Specialty Society
Society of Thoracic Surgeons - Medical Specialty Society

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the American College of Cardiology (ACC) Web site; electronic copies are also available in PDF from the American Heart Association (AHA) Web site.

Print copies: Available from the American College of Cardiology, Resource Center, 9111 Old Georgetown Rd, Bethesda, MD 20814-1699; (800) 253-4636 (US only).

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: executive summary. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol, 2008; 52:1890-1947. Electronic copies: Available from the American College of Cardiology (ACC) Web site. Also available in Portable Document Format (PDF) from the American Heart Association (AHA) Web site.
- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. Slide set. 2008. 88 p. Electronic copies: Available from the American College of Cardiology (ACC) Web site.
- Methodology manual for ACC/AHA Guideline Writing Committees. Methodologies and policies from the ACC/AHA Task Force on Practice Guidelines. 2006 Jun. 61 p. Electronic copies: Available in PDF from the American College of Cardiology (ACC) Web site.

Print copies: Available from the American College of Cardiology, 9111 Old Georgetown Road, Bethesda, Maryland 20814-1699.

PATIENT RESOURCES

None available

NGC STATUS

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